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ORIGINAL ARTICLE

Accelerated myelination along fiber tracts in patients with hemimegalencephaly

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KEYWORDS
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Summary
Background and purpose: In infants with hemimegalencephaly, asymmetrical white-matter intensities suggestive of advanced myelination are observed as well as aberrant midsagittal fibers (AMFs) specific to hemimegalencephaly. Also noted are otherwise unreported abnormally enlarged periventricular fibers (APVFs) running anteroposteriorly along the caudate nucleus. This study investigated the degree of myelination and presence of aberrant fibers in hemimegalencephaly through a retrospective review of MRI scans in relation to histopathological findings.
Materials and methods: MRI scans of 24 infants with hemimegalencephaly (13 boys and 11 girls, 1—9 months old) were evaluated, focusing on the presence and signal intensities of AMFs and APVFs. White-matter signal intensities on T1- and T2-weighted imaging of the cerebral hemisphere were also evaluated and compared with the timetable for normal myelination. Surgical specimens were pathologically examined with Klüver—Barrera staining in four patients.
Results: AMFs and APVFs were observed in 18 and nine patients, respectively, while 22 patients had accelerated myelination of the megalencephalic hemisphere that tended to extend along fiber pathways including AMFs and APVFs. In six cases, accelerated myelination even extended into the contralateral hemisphere via the corpus callosum or AMFs. Histopathological analysis identified hypermyelination with disarrayed myelinated fibers corresponding to MRI findings.

Abbreviations: AMF, aberrant midsagittal fiber; APVF, abnormally enlarged periventricular fiber; FA, fractional anisotropy; PCW, post-conceptional week; SFO, superior fronto-occipital.

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Conclusion: Accelerated myelination is frequently observed in patients with hemimegalencephaly and tends to extend along fiber pathways, including aberrant or abnormal fibers, as seen in 75% of hemimegalencephaly patients. Accelerated myelination may reflect propagation pathways of abnormal brain activity in such patients.

Introduction

Hemimegalencephaly is a rare and unique malformation characterized by enlargement of all or part of a cerebral hemisphere [1,2]. It is considered a heterogeneous malformation that includes several different disorders although its exact pathogenesis is still not completely understood [3]. Magnetic resonance imaging (MRI) findings in hemimegalencephaly include enlargement of the involved cerebral hemisphere, abnormal gyral patterns with thickened cortices, loss of gray—white-matter differentiation and white-matter gliosis [4,5]. Structural changes have also been reported beyond the involved cerebral hemisphere, including ipsilateral olfactory nerve enlargement, cerebrovascular dilation and cerebellar enlargement with abnormal folia [6]. The presence of aberrant midsagittal fibers (AMFs) as a band-like structure lying between the anterior horns of the lateral ventricles, and running beneath the body of the corpus callosum, has also been reported in patients with hemimegalencephaly [7]. Tractography using diffusion tensor imaging (DTI) is a useful tool for visualizing such abnormally located neuronal fiber tracts [8,9], and has successfully demonstrated the course of AMFs penetrating the band-like structure and often passing into the ipsilateral and/or contralateral fornices [7]. In addition, previously unreported abnormally enlarged periventricular fibers (APVFs), running anteroposteriorly between the lateral ventricle and internal capsule, have recently been observed in a hemimegalencephaly patient.

Hyperintensities on T1-weighted imaging (T1WI) and hypointensities on T2-weighted inaging (T2WI) in the white-matter of the affected hemisphere have also been described in a limited number of hemimegalencephaly patients [4,10—12]. Although three of these previous reports suggested that such signal intensities are due to accelerated myelination [10—12], histopathological confirmation has been lacking. Moreover, the previous studies included only small numbers of patients < 2 years old, and their evaluations of white-matter signal-intensity were somewhat ambiguous.

For this reason, the present study aimed to investigate the degree of myelination in infants with hemimegalencephaly through a retrospective review of MRI scans with reference to histopathological findings. The presence and signal-intensity pattern of APVFs as well as AMFs were also determined.

Materials and methods

Patients

A search through our institutional radiological database identified 24 consecutive hemimegalencephaly patients aged < 2 years (13 boys and 11 girls; age: 3.75 ± 2.07 months, range: 1—9 months), and hospitalized at our institution between November 2001 and August 2012. These patients were all full-term at birth except for one preterm infant who was born during week 35 of gestation. All of these infants had been hospitalized for intractable epilepsy to determine their indications for surgical treatment. The diagnosis of hemimegalencephaly was made on the basis of both clinical and imaging findings. The standard evaluation included a detailed clinical history and neurological examination, scalp electroencephalography (EEG) recordings, 99 m-technetium-ethyl-cysteinate-dimer single-photon emission computed tomography (SPECT) and MRI. Twenty-three of our selected patients had undergone functional disconnection surgery, and four had a suitable amount of resected specimens for detailed pathological analyses. The local institutional review board did not require written informed consent for this retrospective review.

MR imaging

Images were obtained using a 1 Tesla (T), 1.5 T or 3 T MRI scanner system. Sequences included axial spin-echo (SE) T1WI (TR, 580—624 ms; TE, 9—15 ms; section thickness, 3—5 mm; intersection gap, 3.3—5.5 mm; matrix, 240—256 × 169—209; NEX, 1 or 2), axial fast spin-echo (FSE) T2WI (TR, 3600—5000 ms; TE, 80—98 ms; section thickness, 3—5 mm; intersection gap, 3.3—5.5 mm; matrix, 368—512 × 263—348; NEX, 1 or 2), axial and/or coronal fluid-attenuated inversion-recovery (FLAIR) images (TR, 8500—12,000 ms; TE, 94—120 ms; TI, 2500—2700 ms; section thickness, 3—5 mm; intersection gap, 3.3—5.5 mm; matrix, 256—320 × 162 — 202; NEX, 1 or 2) and coronal short inversion-time inversion-recovery (STIR) images (TR, 4000—6190; TE, 80—96; TI, 150—230; section thickness, 3 mm; intersection gap, 3.6—4.5 mm; matrix, 312—512 × 215 — 284; NEX, 1 or 2). Sagittal three-dimensional (3D) T1WI was also acquired using either a 3D turbo field-echo (TFE) sequence (TR, 7.3 ms; TE, 3.5 ms; sagittal thickness sections, 1.2 mm; matrix, 320 × 285; NEX, 1) or an inversion-recovery gradient-echo sequence (TR, 1600—2080 ms; TE, 2.6—3.9 ms; TI, 800—1100 ms; sagittal thickness sections, 0.8—1.2 mm; matrix, 256—320 × 208—288; NEX, 1), and reconstructed in axial and coronal planes.

DTI data were available for seven patients (aged 1—5 months). Diffusion was measured along 12 or 15 non-collinear directions with the use of a b factor: 700 s/mm^2 with the 1 T system and 1000 s/mm^2 with the 3 T system. Other parameters were TR, 5760—10,100 ms; TE, 62—113 ms; FOV, 230 × 230 or 240 × 240 mm^2; matrix, 76 × 76 or 80 × 78; section thickness, 3 mm; no intersection gap; and NEX, 5 for the 1 T system and 2 for the 3 T system.